PEDIATRIC TRACHEAL STENOSIS AND VASCULAR RINGS

ÇOCUKLARDA TRAKEAL STENOZ VE VASKÜLER HALKALAR

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Abstract

Pediatric tracheal stenosis includes a variety of entities characterized by a diminished airway lumen. Congenital entities can be classified into (i) intrinsic disease caused by short- (e.g. tracheal webs) or long-segment forms of tracheal stenosis (e.g. tracheal atresia/agenesis and primary tracheomalacia) and (ii) extrinsic disease caused by cardiovascular anomalies such as vascular rings and/or congenital tumors. Acquired entities include intubation trauma and tracheostomy-induced formation of granulation tissue. Surgical and endoscopical techniques developed in the last 20 years have dramatically improved the outcome of these patients. With the wider availability of higher-resolution imaging, minor or subclinical forms are increasingly recognized. Asymptomatic mild cases or symptomatic cases with no respiratory embarrassment can be safely managed in an expectant manner. Very symptomatic patients with an intrinsic, short tracheal stenosis generally undergo resection and anastomosis, whereas in those with a long tracheal stenosis a slide tracheoplasty is performed. Patch tracheoplasty is recommended in patients with intrinsic, long tracheal and bronchial stenosis. Endoscopic techniques such as dilation, laser excision or stents play a large and expanding role in managing complications, although a tracheostomy or second open tracheoplasty might be unavoidable. In place of evidence-based recommendations and standards, patient care remains individualized, based on each institution's expertise and past experience. Close coordination between various services in a multidisciplinary approach can optimize the airway treatment and ensure the most favorable long-term outcome for the child.

Key words: Pediatric tracheal stenosis; vascular rings; endoscopy; open tracheoplasty

Özet

Çocuklarda trakeal stenoz daralmış bir solunum yolu boşluğu ile karakterize çeşitli antiteleri içerir. Konjenital antiteler iki sınıfa ayrılabilir: (i) trakeal stenozun kısa- (örn: trakeal ağlar) veya uzun-segment formlarından kaynaklanan intrinsik hastalık (örn: trakeal atrezi/agenezi ve primer trakeomalazi) ve (ii) vasküler halkalar ve/veya konjenital tümörler gibi kardiyovasküler anomali kaynaklı ekstrinsik hastalık. Kazanılmış antiteler; entübasyon travması ve trakeostomi ile indüklenmiş granülasyon dokusu oluşumunu içerir. Son 20 yılda gelişen cerrahi ve endoskopik teknikler bu hastaların akibetini önemli ölçüde iyileştirmiştir. Yüksek çözünürlüklü görüntülemenin daha yaygın bir şekilde ulaşılabilir olması ile minör veya subklinik formlar artan oranda saptanmaktadır. Semptomatik olmayan hafif vakalar veya solunum sıkıntısı olmayan semptomatik vakalar beklendiği gibi güvenli olarak tedavi edilebilir. İntrinsik, kısa trakeal stenozlu aşırı semptomatik hastalarda genellikle rezeksiyon ve anastomoz uygulanırken uzun trakeal stenozu olanlarda slide trakeoplasti uygulanır. İntrinsik, uzun trakeal ve bronşiyal stenozu olan hastalarda yama (patch) trakeoplasti uygulanması önerilmektedir. Trakeostomi veya ikinci açık trakeoplasti kaçınılmaz olabilmekle beraber dilatasyon, lazer eksizyon veya stentler gibi endoskopik teknikler komplikasyonlarla başa çıkmada büyük ve giderek artan bir rol oynamaktadır. Kanıta dayalı öneriler ve standartlar yerine, her kurumun uzmanlığına ve geçmiş tecrübelerine dayanarak hasta bakımı kişiselleştirilmiş olmayı sürdürmektedir. Multidisipliner bir yaklaşımda çeşitli hizmetler arasındaki yakın koordinasyon solunum yolu tedavisini optimize edebilir ve çocuk için en uygun uzun süreli sonuçları garanti edebilir.

Anahtar kelimeler: Çocuklarda trakeal stenoz, vasküler halkalar, endoskopi, açık trakeoplasti

INTRODUCTION

Stenosing airway disease includes a variety of entities, congenital and acquired, ranging from intraluminal obstructions to extrinsic compressions and malacia. Although they are etiopathogenically different, all of them show a diminished airway lumen that may require surgical or endoscopical treatment.

Congenital tracheal stenosis is an infrequent structural, obstructive lesion often associated with other airway or cardiovascular malformations. Its estimated incidence is 1 in 64,500 newborns, and represents only

0.3% to 1% of all laryngotracheal stenosis (1, 2). Most of them are severe in nature. Before the advent of the current surgical techniques, mortality was reported to be as high as 79%, mainly because of acute airway obstruction and the lack of effective medical treatment (3). Tracheal release maneuvers which were developed in the 1950s later extended the resectable length to 25% to 30% of the trachea while the introduction of cardiopulmonary bypass and the Montgomery T-tube (Boston Medical Products, Westborough, Massachusetts) (used as a stent as well as a tracheostomy tube) began to address the associated and most frequent intra- and postoperative complications (4). By 1982, Kimura et al. (5) had introduced the costal cartilage tracheoplasty for long stenotic segments. Their work was followed in 1984 by Idriss et al. (6), who described the pericardial patch tracheoplasty. Problems with granulation formation and prolonged postoperative intubation were addressed when the slide tracheoplasty was proposed in 1989 by Tsang et al. (7) and modified by Grillo (8). The evolution of these surgical techniques has improved outcomes significantly for previously inoperable patients. In addition, better understanding of postoperative care and management of complications has led to improved reported mortality rates ranging from 9% to 21% (9).

EMBRYOLOGY

The laryngotracheal groove or sulcus appears in the proximal foregut at the third week (3 mm embryo, stage 10). The laryngotracheal groove progresses caudad and the lateral ridges progress cephalad to form the primordium of the trachea. The pulmonary primordium appears and bulges ventrally from the foregut. Complete separation of the trachea and esophagus occurs by the sixth week (11 to 14 mm embryo). The tip of the tracheal primordium buds asymmetrically, left and right, at the 4 mm stage, to provide bronchial primordia. Mesenchymal proliferation by cells lining the coelomic cavity provides the tissue from which cartilage, muscle, and connective tissue will develop. Epithelial–mesenchymal interrelationships are essential for bronchial and pulmonary development to occur. The tracheal bifurcation moves gradually downward from the neck to the level of the fourth vertebra. Cartilage appears in the trachea at the tenth week.

When the laryngotracheal groove appears, the forerunner of the glottis also appears as a median slit in the pharyngeal floor between the fourth and sixth branchial arches. The epiglottic primordium lies anteriorly and the arytenoid swellings lie laterally prior to their more medial migration. Ventricular buds are solid at first. A T-shaped slit appears, which opens into a lumen by the eighth week. Vocal cords are seen at 3 months. Thyroid and cricoid cartilages appear between 5 and 7 weeks. The laryngeal cartilages derive from the fourth and fifth arches.

Failure of complete separation of the foregut into respiratory and alimentary components is the most common defect and produces a tracheoesophageal fistula (TEF). At the upper end, the larynx may fail to reopen, producing atresia (a fatal anomaly), or it may fail to form a complete posterior septum, producing a laryngotracheoesophageal cleft. Tracheal atresia, stenosis, esophageal atresia, and tracheoesophageal fistula occur more distally. The relatively separate processes of laryngeal development and budding of bronchi and pulmonary development allow for malformations of the trachea, such as agenesis and stenosis in the presence of a normal larynx and bronchial tree (10).

CLASSIFICATION

Multiple classification schemes for tracheal stenosis have emerged, perhaps because of the spectrum of causes, manifestations and disease severity. Definitions have attempted to encompass morphology, severity and mortality. Tracheal stenosis can generally be divided into acquired versus congenital forms, extrinsic versus intrinsic disease, and short-segment versus long segment stenosis.

Congenital forms of tracheal stenosis were first classified by Cantrell and Guild (11) (Figure 1), who categorized the condition as generalized hypoplasia, funnel shaped stenosis, and segmental stenosis. The stenotic segment is most often composed of completely circular “O” rings of cartilage. Alternatively, disorganized cartilages, ridges, or plates of cartilages may occur. Hoffer et al. (3) subsequently accounted for the length of stenosis as well as associated anomalies, both factors that affect mortality and treatment. Class 1 is described as short-segment stenosis with the best prognosis (8% mortality), class 2 as long-segment stenosis with anomalies other than pulmonary or cardiovascular malformations (45% mortality), and class 3 as any stenosis with pulmonary or cardiovascular malformations (79% mortality).

Acquired forms of pediatric tracheal stenosis include intubation trauma and tracheostomy-induced formation of granulation tissue. Injury from endotracheal tubes is the most common cause of tracheal

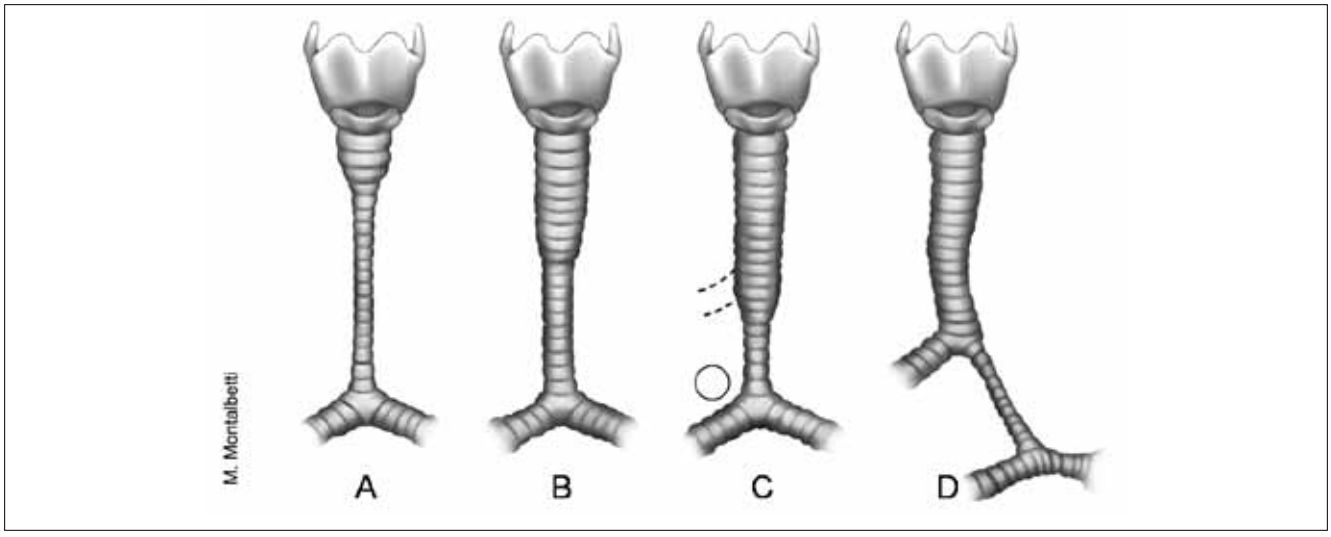

Figure 1. General categories of congenital tracheal stenosis. Types I–III redrawn from Cantrell JR and Guild HC (10,11); A) Type I: Long segmental stenosis, all or most of the trachea is stenosed. B) Type II: Funnel shaped stenosis with variable tracheal length and location. C) Type III: Short segmental stenosis, sometimes below an anomalous right upper lobe bronchus. D) Type IV: Anomalous right upper lobe bronchus with a "bronchus" to horizontally branching bronchi to the rest of the lung. The right upper lobe bronchus is at the normal carinal level. The bridge bronchus is stenotic, and lesser stenosis may involve part of the trachea above. In some cases, the trachea is elongated as shown, but the upper lobe bronchus is absent. Circles indicate locations of left pulmonary artery sling when present

stenosis and results from cuff pressures greater than the mean capillary pressure of the tracheal mucosa (20 mm Hg). Occlusion of the blood supply facilitates erosion and distortion of the tracheal architecture, leading to fibrous maturation of a lumen stricture. Such a stricture may develop after only 36 hours of intubation. Symptoms are usually apparent 5 weeks after intubation (12). The introduction of compliant, high-volume, low-pressure cuffs has reduced the danger of endotracheal tubes significantly, with the incidence of stricture dropping from as high as 20% of all intubations to current reports of between 1% and 8% of all neonates who undergo prolonged intubation (12).

1. Tracheal Webs

Tracheal webs represent an intrinsic, short-segment form of tracheal stenosis. The web is a tissue layer of variable thickness draped circumferentially around the airway lumen. The underlying cartilage framework is generally intact (13). Webs may be generally managed endoscopically with laser treatment or cutting instruments. Firmer, more involved strictures may require an open resection and re-anastomosis.

2. Tracheal Atresia/Agenesis

Tracheal atresia/agenesis is an intrinsic, long-segment form of congenital tracheal stenosis characterized by partial (tracheal atresia) or complete absence of the airway (tracheal agenesis) (Figure 2) (14). The incidence is 1:50,000; 52% of patients are premature, and up to 94% have other congenital malformations (15). It is usually fatal at birth. The larynx may form normally. The lungs may or may not be normal, and with or without bronchial communications to the esophagus. The most common presentation is with normal bronchi, communicating centrally to the esophagus. Other congenital anomalies are common in these latter patients. Fonkalsrud et al. (16) described a newborn of type C, who survived for a short term by using the esophagus as an airway. A major bronchus may also communicate directly with the esophagus, while the balance of the lung is served by anomalous bronchi from a partly stenotic trachea. Microgastria is a common concomitant feature. No systematic surgical treatment has evolved, doubtlessly due to the rarity and variations in the anomalies as well as the complexity of the defects.

Hiyama et al. (17) described two such patients in whom diagnosis was suspected due to respiratory distress without audible cry and difficulty in intubation. One infant was successfully treated by the following procedures: gastrostomy and abdominal esophageal banding, translaryngeal and esophageal ventilation by endotracheal tube, tracheostomy and later T tube, pharyngeal sump drainage followed by establishment of cervical esophagostomy (proximal tracheal segment present), and esophageal reconstruction by colonic interposition at age 3.

3. Tracheomalacia

Primary tracheomalacia is an intrinsic, long-segment variant of tracheal stenosis characterized by weakened integrity of the tracheal wall caused by insufficient cartilaginous support. The resulting flaccidity results in widening of the posterior membranous wall, with the cartilage-to-membranous wall ratio falling from 4.5:1 to 2:1 to narrow the airway. The posterior wall advances anteriorly during expiration to narrow the lumen even more, resulting in expiratory stridor, brassy cough, neck hyperextension, and reflex apnea in severe cases. The condition may occur in premature as well as full-term infants and may be associated with laryngomalacia (13). Diagnosis is made via endoscopy in a spontaneously breathing patient, because positive pressure ventilation interrupts any dynamic collapse. Primary tracheomalacia is typically self limited to the second year of life; treatment involves observation, positioning, control of reflux, and bilevel positive airway pressure until the patient outgrows the condition. Severe cases may require tracheostomy or stent placement.

Focal tracheomalacia may also occur in relation to a widened membranous wall or residual pouch after repair of TEF and esophageal atresia. Defective cartilaginous rings may also be found at this level. Filler et al. (18) considered this association to be the most common cause of tracheomalacia in infants, although a precise explanation is lacking. If severe collapse follows TEF repair, aortopexy may have to be considered (19). Since gastroesophageal reflux can also be present, antireflux surgery may be needed.

4. Vascular rings

Vascular rings are extrinsic, short-segment forms of congenital tracheal stenosis. The complexity of embryologic development between the cardiovasculature and the airway can easily result in compression involving the trachea, esophagus or both. Up to 50% of the patients who have congenital tracheal stenosis also have such anomalies (20).

Innominate artery compression is perhaps the most common vascular anomaly, resulting from abnormally distal or midline take-off from the aorta. Bronchoscopy typically reveals pulsatile anterior compression of the trachea without esophageal compression (Figure 3). Patients may present with apnea, expiratory stridor, or recurrent pneumonitis (21). Management typically is observation, with inominopexy or aortopexy to the sternum reserved for significant persistent symptoms.

A double aortic arch (complete vascular ring) requires surgical intervention (22, 23). The malformation compresses both the trachea and esophagus together in a circumferential fashion, with either the anterior or posterior arch around the larger vessel (Figure 4). Dysphagia, as well as dyspnea, is usually present, and barium swallow typically demonstrates posterior esophageal compression. However, vessels are now identified by computed tomography (CT) with contrast or by magnetic resonance imaging (MRI). Definitive management involves division of the ring by ligating the smaller aortic arch.

Segmental stenosis of the distal trachea may be associated with an aberrant left pulmonary artery, the

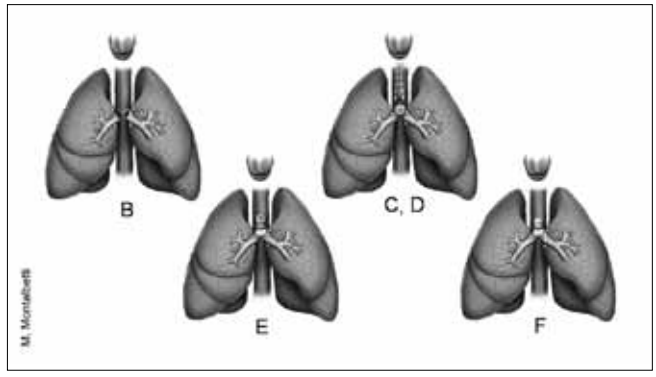

Figure 2. Tracheal agenesis, redrawn according to Faro and to Grillo´s classification (10). A denotes total pulmonary agenesis. In type B, both main bronchi connect to esophagus separately, whereas in type C, bronchi are fused and a bronchoesophageal fistula (BEF) is present. Type D additionally has an atretic band (dashed line) from the larynx to BEF. Type E has a tracheoesophageal communication, whereas in type F, communication with the esophagus is absent. Type G shows short segmental tracheal agenesis

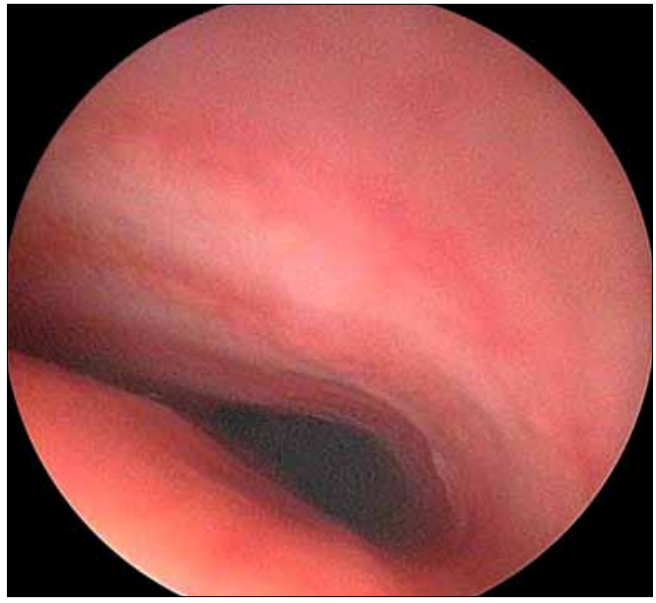

Figure 3. Bronchoscopic image of innominate artery compression

so-called “pulmonary artery sling” (Figure 5) (24, 25). The left pulmonary artery originates from the proximal portion of the right artery and passes behind the trachea to the left lung. In most of these patients, completely circular “O” rings of cartilage are found in the stenotic segment. The length of tracheal stenosis most often extends beyond the region of the anomalous pulmonary artery sling.

Division of the anomalous pulmonary artery and reimplantation into the main pulmonary artery anterior to the trachea fails to relieve the airway obstruction when stenosis or malacia are present (25, 26). The ligamentum arteriosum, which effectively makes this a ring, is also divided. Sometimes, it has been possible to resect the stenotic tracheal segment and shift the artery anteriorly prior to anastomosing the trachea, as suggested might be possible by Grillo (27, 28). This is desirable because of a high rate of stenosis of reimplanted pulmonary arteries in children. The anatomic disposition of the pulmonary arterial sling, however, does not generally permit this transposition without division and reimplantation of the anomalous artery. The distortion of the artery may also compress the resected trachea and cause recurrent obstruction (29).

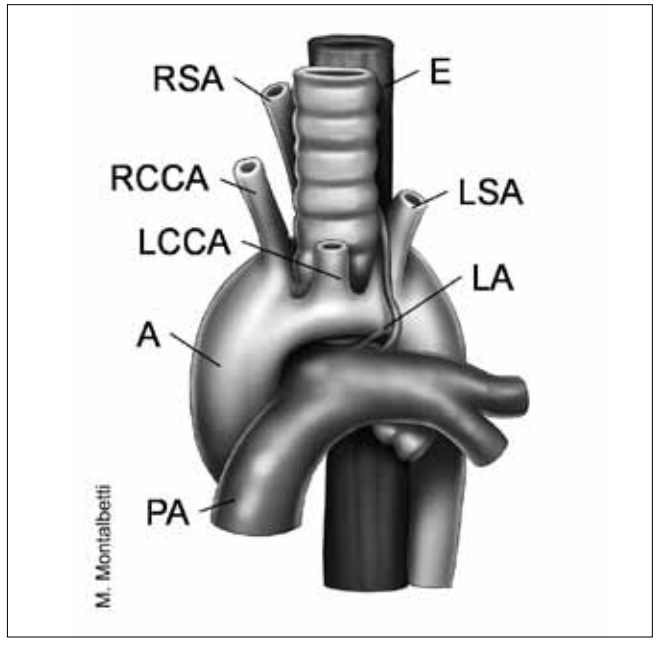

Figure 4. Mayo Type IA: double aortic arch, dominant right arch. Adapted from Stewart et al (23)
A: aorta; E: esophagus; LA: ligamentum arteriosum; PA: pulmonary artery; RCC, LCC: right, left common carotid arteries; RS, LS: right, left subclavian arteries; V: vagus nerve

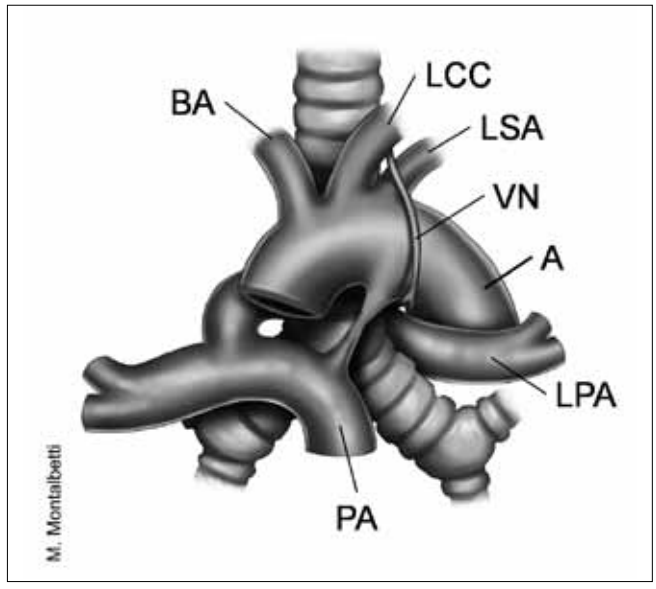

Figure 5. Anatomic relationships with anomalous left pulmonary artery sling. Anterior view shows the aortic arch and ligamentum arteriosum. Note position of vagus and recurrent laryngeal nerve
A: aorta; BA: brachiocephalic artery; LCC: left common carotid; LPA: left pulmonary artery; LSA: left subclavian artery; PA: pulmonary artery; VN: vagus nerve

CLINICAL FINDINGS

The diagnosis of congenital tracheal stenosis and other obstructive anomalies is based on a high degree of suspicion in infants and children with respiratory distress. Tracheal stenosis classically presents as biphasic stridor within the first few weeks of birth. The inspiratory stridor results from stenosis in the cervical trachea, whereas the expiratory stridor results from narrowing or collapse in the thoracic trachea. Associated symptoms include a brassy nonproductive cough, ‘‘washer machine’’breathing, nasal flaring, wheezing, intercostal retractions, pursed lips, persistent croup, and intermittent cyanosis. The infant may hyperextend the neck to straighten the narrowed airway or fail to thrive because of the increased work of breathing and eating at the same time. Importantly, only a 1-mm decrease in an infant’s airway results in a 44% decrease in cross-sectional area. Symptoms may not become apparent until 50% stenosis occurs; dyspnea at rest is likely to present at 75% stenosis (30). Accordingly, presentation often is delayed until the infant develops an upper respiratory infection that exacerbates the narrowed lumen, leading to reflex apnea or dying spells.

Acquired stenosis due to intubation for ventilation is signaled by shortness of breath on exertion and stridor in the wake of a history of intubation, usually for respiratory support, with or without tracheostomy. Children diagnosed with “asthma,” who fail to respond to treatment, must be suspected of an organic airway lesion. If the child has been previously ventilated, the working diagnosis should be airway stenosis, until proved otherwise.

DIAGNOSTIC IMAGING

Air tracheograms and careful fluoroscopy provide precise information about the presence of a tracheal anomaly and its location (Figure 6). CT scanning provides precise information on the cross-sectional area and extent of lesions. An advantage of spiral CT is the rapidity of examination, which is important in small children. MRI offers similar information and is especially useful to delineate associated cardiovascular anomalies. Angiography is also used less often, but it still provides a "gold standard" for precise and complete delineation of vascular anomalies. Echocardiography is also very helpful to identify cardiovascular anomalies. Barium swallow is still an important diagnostic method for identifying a vascular ring or aberrant subclavian artery (Figure 7).

Virtual endoscopy is created from helical CT reconstructions of the airway´s inner surface, and can document stenotic areas where a bronchoscope cannot pass through (Figure 8). As a noninvasive technique, it is useful for patients unable to tolerate bronchoscopy and has been shown to be as accurate as fiberoptic bronchoscopy for central airway stenosis, with reported accuracy rates of 95.5% (31, 32). Detection is poor at the segmental airway level, however. It has a current maximal spatial resolution of 1.5 mm, causing it to miss subtle stenotic lesions. It does not depict tracheal mucosa and it cannot image the airway dynamically.

ENDOSCOPY

Careful use of a flexible pediatric bronchoscope can clarify much about a lesion. The bronchoscope should not be passed into a tightly stenotic lesion in order to avoid causing edema and inflammation, which might precipitate acute obstruction. Indeed, in a significantly symptomatic child, in whom the presence of stenosis is already known from a radiologic study, bronchoscopy is usually best deferred to the time of planned surgical repair. This also applies to less critical patients.

Definitive bronchoscopy, ideally performed just prior to a planned surgical procedure for correction of

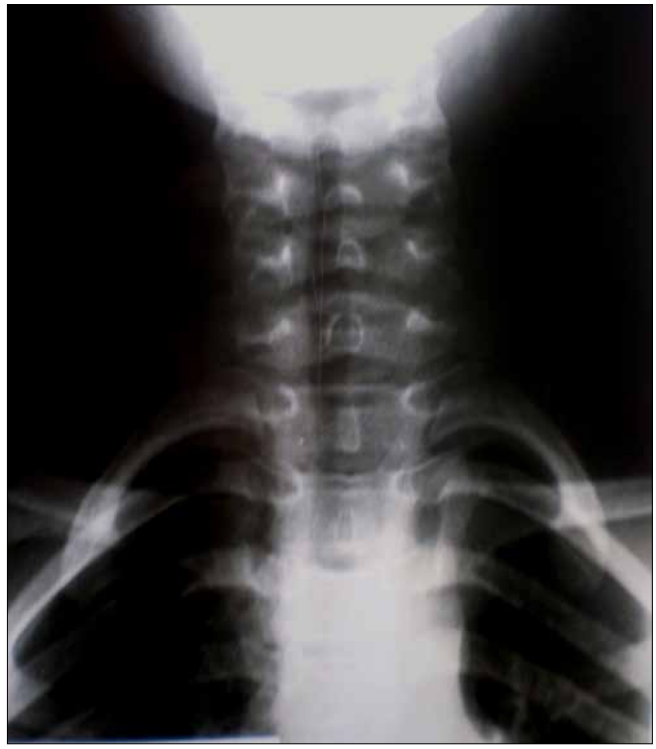

Figure 6. Air tracheogram shows tracheal lumen reduction in the cervico-thoracic junction

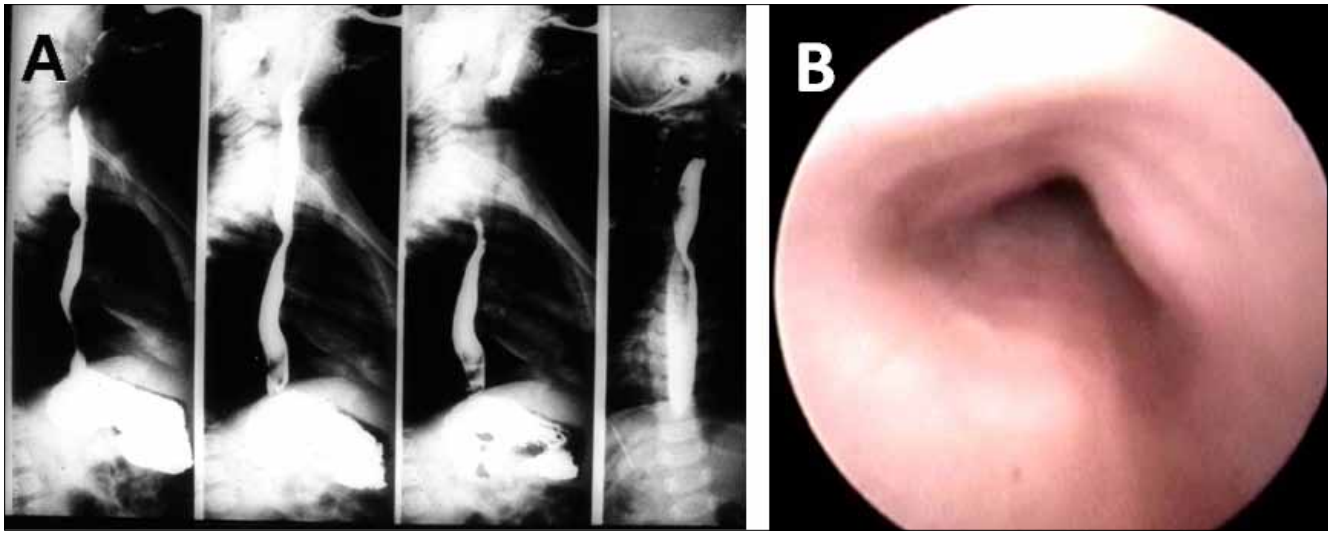

Figure 7. A) Barium swallow in patients with double aortic arch. B) Tracheoscopic image shows narrowing of the anterior and lateral esophageal lumen

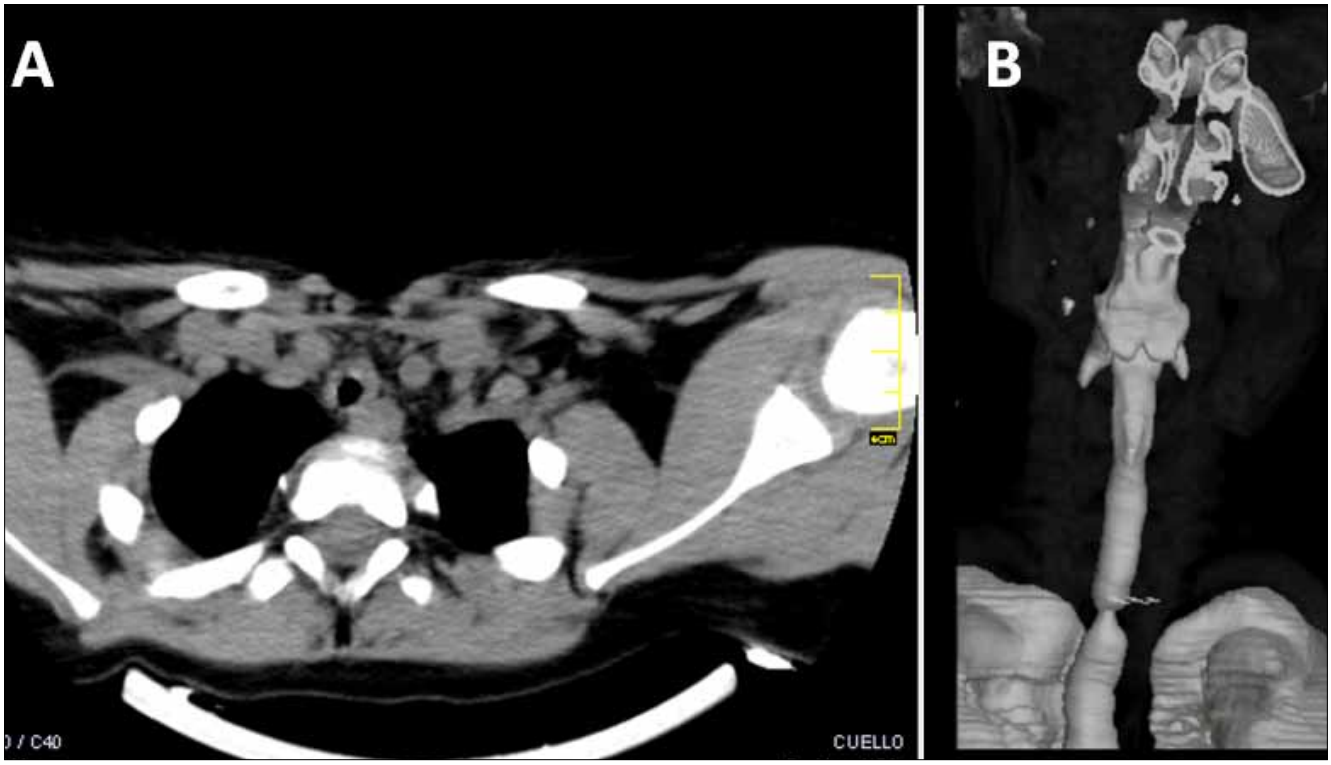

Figure 8. A) CT cross-sectional area of compromised trachea. B) Helical CT reconstructions of the tracheal inner surface

the lesion (Figure 9), is best accomplished with rigid Storz ventilating pediatric bronchoscopes. The 3.5 mm rigid bronchoscope (OD 5.7 mm) will not pass through a tiny stenosis. However, either a flexible pediatric bronchoscope (2.7 mm) or a long telescope (OD 2 mm) allows a more distal examination. These may be inserted through a larger rigid (ventilating) bronchoscope located proximally, or through a pediatric laryngoscope. As in adults, the larynx must be examined, especially in the presence of a postintubation lesion. The glottis is visualized in passing with the rigid bronchoscope and telescope. However, the pediatric Holinger laryngoscope, used in conjunction with a telescope, provides a superior view of laryngeal structures (33, 34).

TREATMENT

Surgical and endoscopical techniques developed in the last 20 years have dramatically improved the outcome of these patients. Currently, the challenge lies more in reducing the high morbidity rate associated with therapeutic procedures rather than in achieving survival. Nevertheless, not every patient with tracheal stenosis requires operative treatment; asymptomatic cases can be managed in an expectant manner and eventually may not need surgical treatment (35). Greater understanding of the disease has also led to better stratification of management based on symptoms (Figure 10).

1. Conservative Management

With the wider availability of higher-resolution imaging, minor or subclinical forms of tracheal stenosis are increasingly recognized. Although most patients who have tracheal stenosis require intervention, it has been shown that a subset may be observed instead (36, 37). The expense and risk of high morbidity and mortality of tracheoplasty have made observation a more palatable option. Medical management includes antireflux treatment, antibiotics for infection, chest physiotherapy, and humidified air.

One longitudinal study following six patients for an average of 10.6 years with CT scans found that stenotic tracheas naturally display catch-up growth, with the trend suggesting that patients regain normal lumen diameters by 9 years of age (36). In another study, Rutter et al. (37) followed 10 patients who had complete tracheal rings for at least 12 months. Five patients demonstrated tracheal growth and, accordingly, resolution of symptoms; two required tracheoplasty, and three were too young for the final outcome to be known. Based on these data, it was hypothesized that up to 10% of patients who have complete tracheal rings will not require surgery. Observation is a safe, viable approach for clinically mild tracheal

stenosis to determine if an operation will eventually be needed. Delay is also advantageous, because tracheoplasty is easier to perform on an older, larger infant (37).

2. Endoscopic Management

Endoscopic treatments include laser excision, dilation, and stent placement. Topical placement of steroids or mitomycin C is commonly used in conjunction

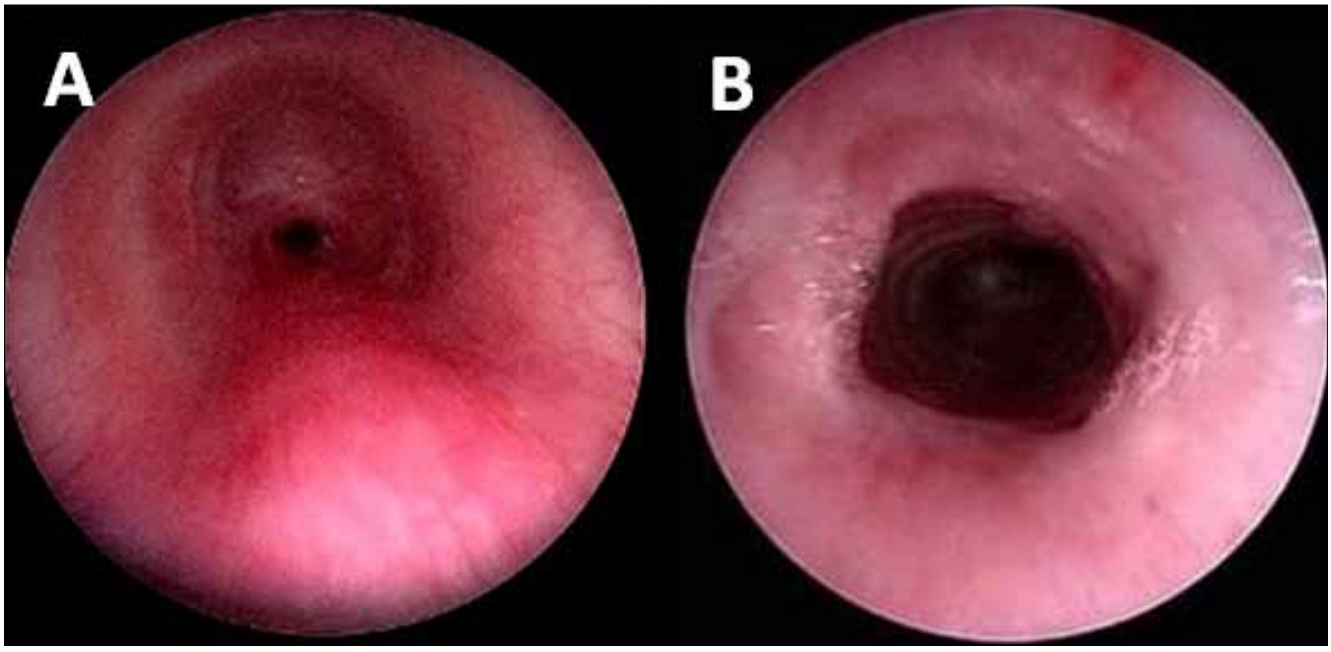

Figure 9. A) Bronchoscopic image before surgery showing the presence of a severe, acquired tracheal stenosis. B) Same patient´s bronchoscopic image after surgery

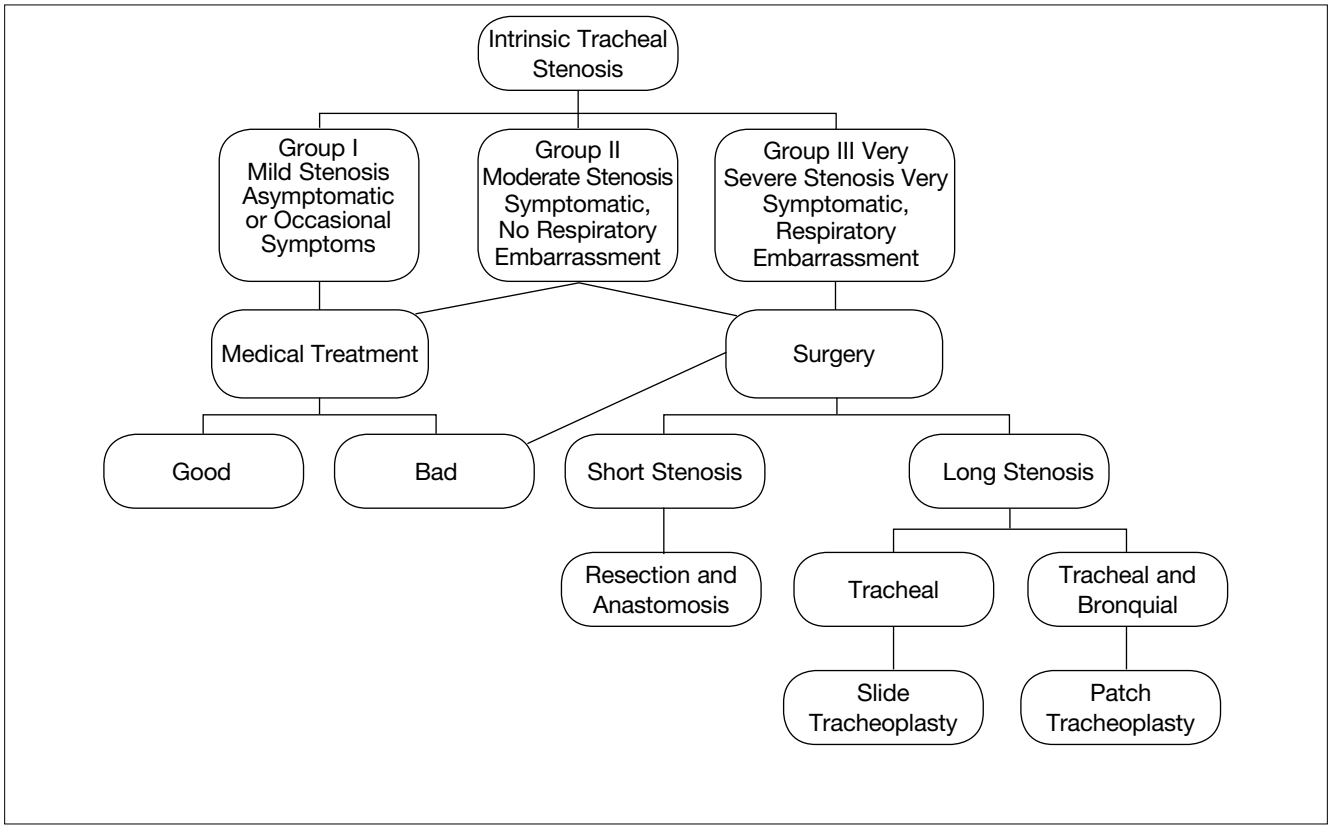

Figure 10. Intrinsic tracheal stenosis: management algorithm

with these approaches. Tracheal webs and tracheomalacia can be treated primarily using endoscopic techniques, with excellent results.

The CO_2 laser is preferred to the neodymium-doped yttrium aluminum garnet (Nd:Yag) or potassium titanyl phosphate options in the trachea because of its limited thermal effect, but the Nd:Yag laser is favored for managing hemostasis (38). Optimal approaches include creating a micro trapdoor or a radial incision followed by dilation. Essential principles of laser work include avoiding cicatricial (scarring), carinal involvement, and compromise of cartilaginous support (21). Lesions more than 1 to 2 cm thick in vertical length normally are considered inappropriate for laser excision.

Balloon techniques localize distending forces in an outward radial direction while avoiding the longitudinal shear forces created by traditional metallic dilators. Trauma to the larynx and tracheal mucosa is minimized, and the dilator can be moved easily to multiple positions. In addition to palliation, Philippart et al. (39) reported the use of balloon dilation to avoid the need for redo open tracheoplasty on patients previously treated with patch tracheoplasty. Despite its advantages, balloon dilation carries significant risks. Much like laser excision, repeat balloon dilations are usually -required to achieve a sufficiently wide airway. Complications including tracheitis, pneumomediastinum, tracheal laceration or perforation, stent displacement, and death from bronchial leak also have been reported (39). The long-term effectiveness of balloon dilation, with or without stent placement, remains unclear for tracheal stenosis and merits further study.

Permanent stents of small caliber or expandable stents should be avoided, because of their failure to account for growth, their ease of occlusion, their potential to cause additional injury, and the difficulty of removal (Figure 11). Congenital segmental stenosis should not be dilated since this can split the “O” rings, which are usually present.

3. Open Surgical Management

The evolution of surgical techniques has enabled far better management of congenital or acquired tracheal stenosis, including the most life-threatening forms such as long-segment stenosis. Cardiopulmonary bypass and ECMO have helped immensely in stabilizing the patient intraoperatively during repair. Coexisting cardiovascular malformations such as pulmonary artery slings are usually repaired concurrently. Postoperative management and surveillance for complications such as granuloma formation is essential, as is a strong commitment from multiple disciplines to monitor and optimize an airway that may remain tenuous for years.

4. Resection and Anastomosis

Increasingly, tracheal resection and anastomosis, both for acquired lesions and short congenital stenoses, has been performed with considerable success.

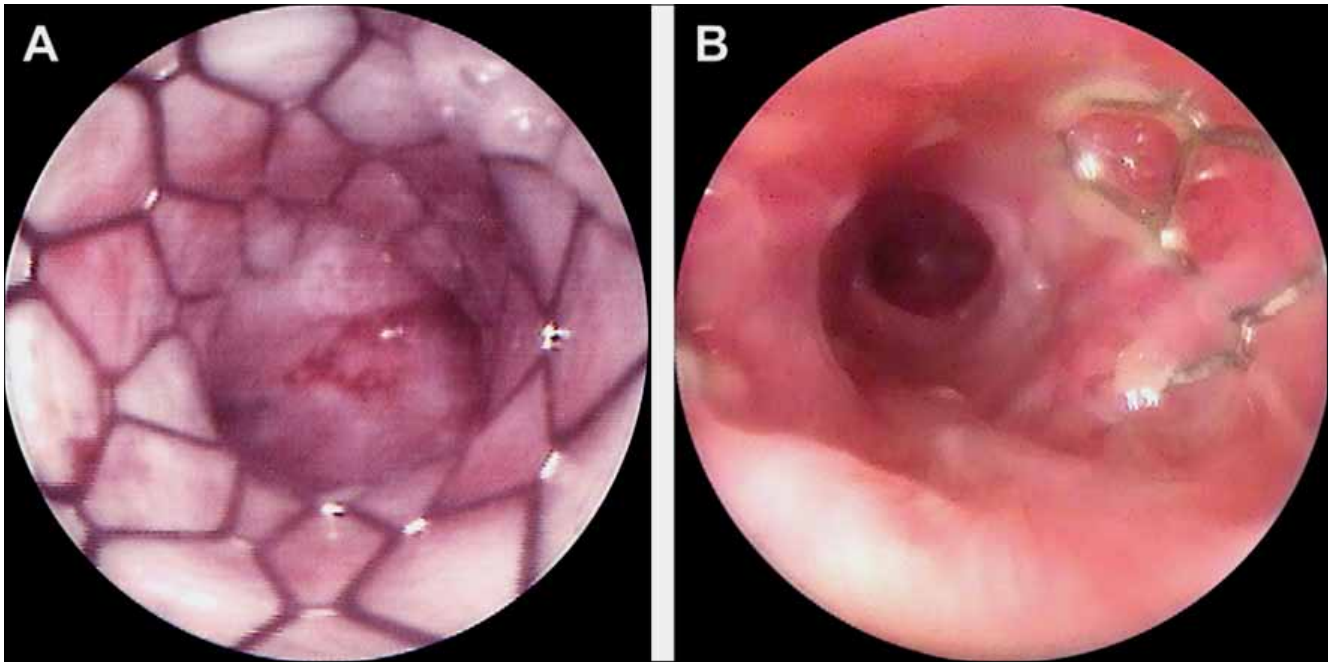

Figure 11. A) Distal tracheal stent placed correctly above the carina in a 3-month-old patient. B) Stent buried in granulation tissues 1 year later

Up to 30% of the trachea may be removed safely, with both proximal and distal portions mobilized circumferentially (Figure 12). The use of lateral traction sutures to reduce tension on the suture line lessens postoperative narrowing (Figure 13). Procedures developed for adults have served well in children, but principally for acquired stenosis, since the length of most congenital stenoses precludes resection and reconstruction. Warnings about intolerance of greater anastomotic tension and danger of postoperative obstruction due to edema and secretions remain valid. Technique, as ever, must be precise and meticulous.

Vicryl sutures have been shown to be as close to an ideal tracheal anastomotic suture as has yet been offered, in terms of ease of use, strength, minimal reactivity and, most importantly, absence of long-term complications such as granulomas, suture erosion into the lumen, and anastomotic separation and stenosis (40). PDS is somewhat more difficult to handle and has no positive advantages to highlight its recommendation.

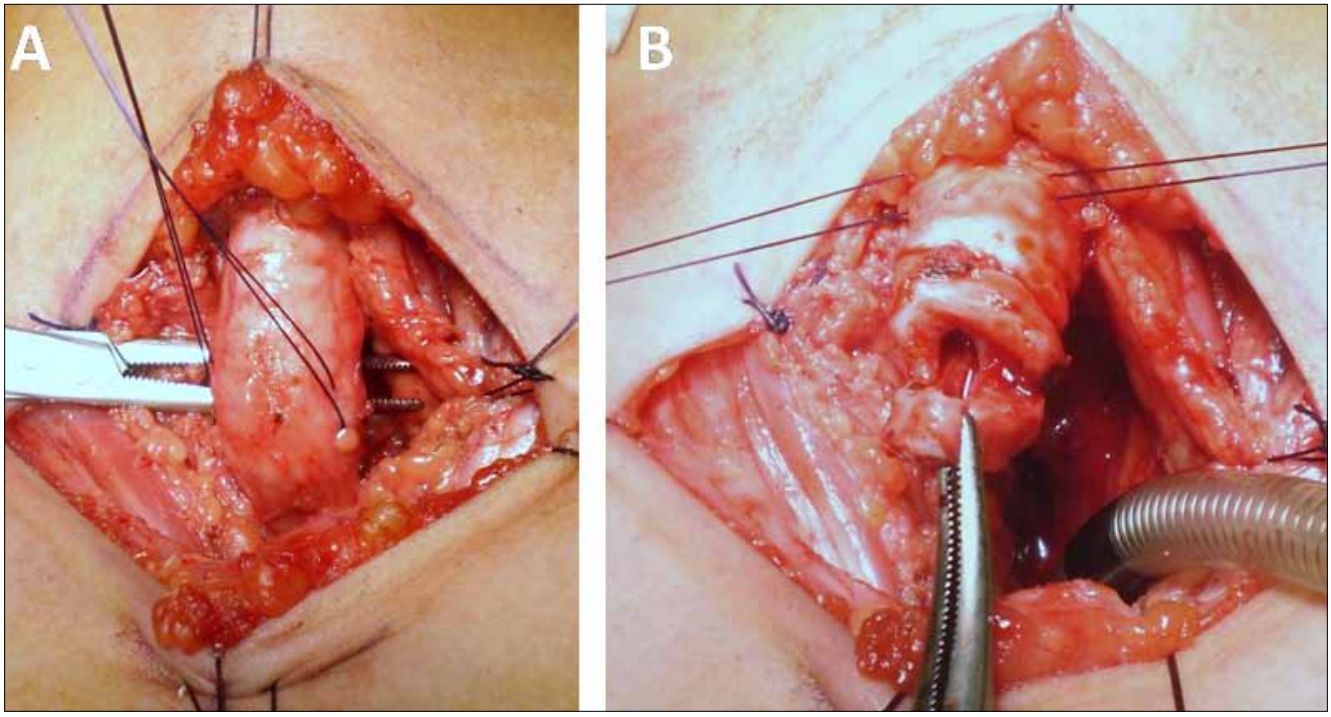

Figure 12. A) Circumferential dissection for B) segmental resection at the site of tracheal stenosis

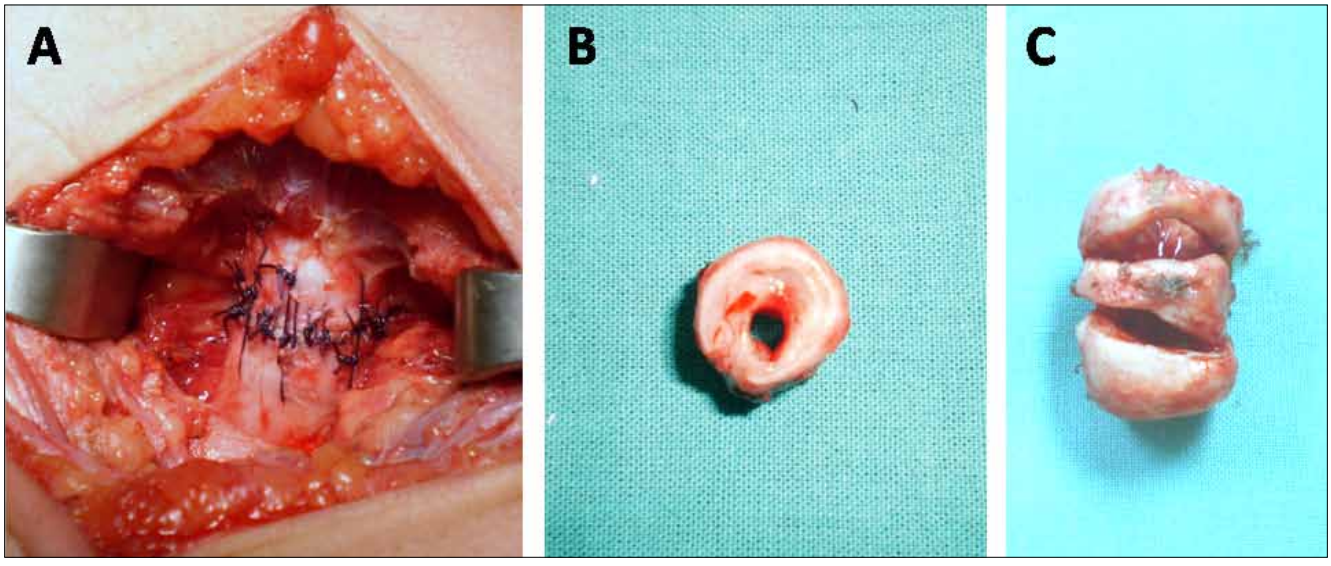

Figure 13. A) The use of lateral traction sutures to reduce tension on the suture line diminished postoperative narrowing B, C) Resected tracheal segments

Grillo et al. (41) reported on resection and anastomosis in 503 adult and pediatric patients who had postintubation tracheal stenosis, finding 87.5% to be free of symptoms, 5.7% to have dehiscence, and a mortality rate of 2.4%. Other single-center studies have reported mortality ranging from 0% to 16%, suggesting the importance of patient selection (42, 43). Time to extubation and frequency of surveillance bronchoscopy, both to monitor the integrity of the anastomosis, vary by institution, and at our institution extubation is from the immediate postoperative period up to 7 days and endoscopic controls a month and then 3, 6, 12 and 24 months. Short-term complications include leaks, mediastinitis, and granuloma formation. Long-term risks mainly involve restenosis.

5. Slide Tracheoplasty

First proposed by Tsang et al. (44) and successfully employed by Acosta (20), this meets the problems of long segment congenital stenosis by using tracheal tissue alone to widen the lumen, giving stability, minimizing granulation tissue formation, and assuring more likely prompt healing. A complete epithelial surface is also immediately provided. The stenosis is divided horizontally in its midpoint (Figure 14A), and the upper and lower segments of stenosis are incised vertically through their entire extent, one anteriorly and one posteriorly (Figure 14B).

Corners are trimmed and the two segments slid together for suturing (Figure 14C). The circumference of the trachea is doubled and the cross-sectional area approximately quadrupled. Since the ends of the cartilaginous walls tend to curl inward, a lobulated cross-sectional appearance of a Figure 8 character may be produced, so that the area is slightly less than quadrupled (Figure 14D). Since the affected segment of the trachea is halved in length, even stenosis of the entire length of trachea does not preclude the procedure.

Compared with autologous patch tracheoplasty, the duration of postoperative intubation and postoperative hospitalization tends to be shorter and more benign for slide tracheoplasty, probably because a stent is not required for support (8, 20, 45). Mortality, based on a compilation of single-center reports on infants by Elliott et al. (12), was 12.5% and by Rutter et al. (46), was 18%, ranging from 0% to 25% (45). Long-term follow-up of patients by Grillo et al. (4) and others have shown patent airways and postoperative tracheal growth (Figure 15).

6. Patch Tracheoplasty

Despite the effectiveness of the slide tracheoplasty, it is contraindicated in certain anatomic variants of pediatric tracheal stenosis. It should not be used in patients who have any bronchial involvement, including a pig bronchus. Although a variety of substances have been used, the most commonly accepted have been costal cartilage, pericardium, tracheal autograft, and tracheal allograft. Synthetic grafts have generally failed because of high infection rates and excessive granulation formation. A vertical incision is made anteriorly, spanning only the stenosis but including the carina or bronchi if necessary. The patch is then sutured over the anterior gap, broadening the tracheal lumen. Cardiopulmonary bypass is generally used in conjunction with patch tracheoplasty, as are stents if necessary to treat unmasked tracheomalacia or bronchomalacia.

Rib cartilage is the first successful graft developed for tracheoplasty. Its advantages include low donor-

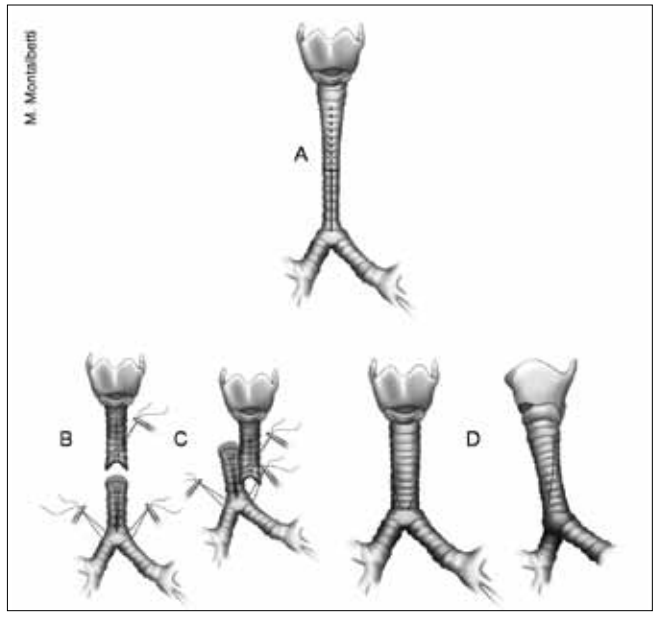

Figure 14. Technique of slide tracheoplasty. The extent of stenosis is precisely defined by intraoperative examination and if necessary by bronchoscopy. The stenotic segment is divided at its midpoint. This is circumferentially dissected in order to preserve as much lateral blood supply as possible. The lower stenotic segment will be divided anteriorly. The upper stenotic segment will be divided posteriorly for the full length of the stenosis. B) The right-angled corners at the midpoint tracheal division are trimmed above and below. A single traction suture is placed near the tip of the upper segment and two lateral sutures below the stenosis inferiorly. Only if absolutely necessary dissection is done, in order to conserve tracheal blood supply. Tracheal circumference will be doubled. C) After all anastomotic sutures have been placed and organized in orderly array, the segments are slid together, with the help of the stay sutures, and the anastomotic sutures are tied. D) An oblique suture line results, with quadrupling of the cross-sectional area, since the circumference of the trachea has been doubled

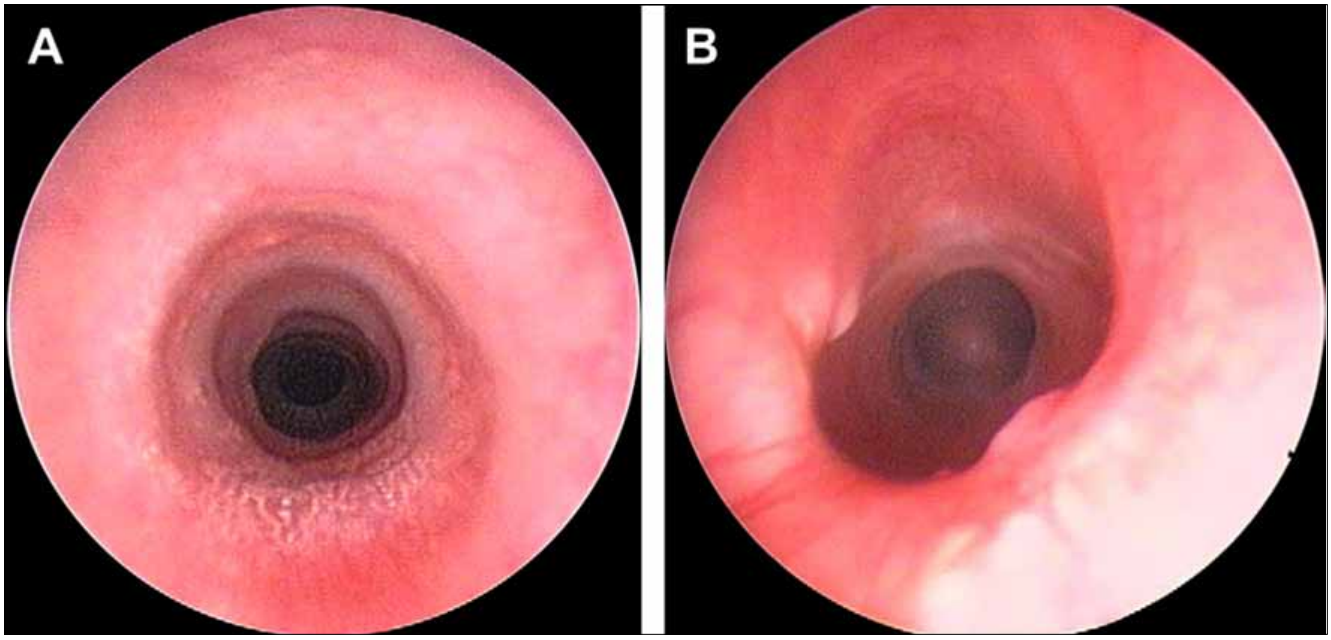

Figure 15. A) Complete tracheal ring before slide tracheoplasty. B) Complete tracheal ring after slide tracheoplasty

site morbidity, plentiful supply, and the ability to survive without requiring a direct blood supply.

COMPLICATIONS

A strong commitment to patient care is imperative in the postoperative period, because the rate of complications continues to be high after open tracheoplasty. Endoscopic techniques such as dilation, laser excision, or stents play a large and expanding role in managing complications, although a tracheostomy or second open tracheoplasty is sometimes unavoidable.

Granulation tissue is perhaps the most common problem seen and risks obstructing an already unstable, narrowed lumen. Formation may occur at the margin between graft and trachea, from exposed suture, from stents, or at the stoma from a tracheostomy. Increased tension also tends to spur aberrant growth of granulation. Therefore tension-dependent techniques such as resection and anastomosis carry a higher risk of granulation. Cold excision, lasers, and progressive balloon dilation have all met with varying amounts of success but risk granulation regrowth or exacerbation. Topical adjunct treatment consists of steroids or mitomycin C, an antimetabolite that inhibits fibroblast formation.

Restenosis of any open tracheoplasty technique is common. The severity of the original stenosis has a major effect on the probability of recurrence (46). Backer et al. (43), reviewing 50 pediatric patients who had complete tracheal rings over 18 years, found that reoperation or stent placement was necessary for 25% of pericardial tracheoplasty patients (7/28), 17% of autograft patients (2/12), 0% of resection and anastomosis patients (0/8), and 50% of slide tracheoplasty patients (1/2). On long-term follow-up of eight pediatric patients who had slide tracheoplasty, Grillo et al. (4) found a rate of restenosis of 0%. Treatment involves bronchoscopy to identify the extent or type of stenosis (firm narrowing or variable collapse), followed by repeated dilation or stent placement.

CONCLUSION

The management of pediatric tracheal stenosis has evolved dramatically in the past several decades and includes surgical measures and treatment for recurrence, complications, and palliation. Conservative management also has been established for milder cases, borne out over a number of years as stenotic tracheas catch up in size. Because of the rarity of the condition, no randomized, prospective, controlled trials yet exist, but they clearly are needed, given the expansive range of therapeutic options and the still unacceptably high rate of complications and mortality. In place of evidence-based recommendations and standards, patient care remains individualized, based on each institution's expertise and past experience. Nonetheless, close coordination between various services in a multidisciplinary approach can optimize the airway and ensure the most favorable long-term outcome for the child.

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